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Independent Study

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Introduction

Hearing is a crucial link in the development of normal speech and language. The human ear has evolved in such a way as to be tuned with sensitivity to the range of human speech (Northern & Downs, 1991). Speech is the primary mode of human communication, but its acquisition appears to rely upon auditory experiences during a critical period between the ages of birth to three years. With deprivation of such auditory experiences, a child's speech and language development will never fully attain its potential (Northern & Downs, 1991). Additionally, delays in speech and language development can cause delays in other aspects of child development, such as literacy, overall academic achievement, and social and emotional development (Hayes & Northern, 1996). Hearing loss during childhood, therefore, can have far-reaching effects on a child's development. These effects on development can be lessened with intervention. However, due to the brief and early critical period for language learning, this intervention must occur as early in the hearing impaired child's life as possible. The invisibility of hearing impairment is part of the reason that the average age for identification of hearing loss is thought to currently be between two and three years of age, nearly the end of the critical period for speech and language learning (Northern & Downs, 1991; Hayes & Northern, 1996). It is necessary to establish a way in which children with hearing loss can be identified at a much earlier age. This has come in the form of infant hearing screening programs.

In 1970 the Joint Committee on Infant Hearing Screening (JCIH) was formed to evaluate the status of infant hearing screening programs. At that time the JCIH included a high risk register, which contained risk factors for hearing loss. The high risk register has been modified repeatedly, most recently in 1994. The JCIH then updated their recommendations as to the best methods of screening infant hearing with the goal of early identification. It has been estimated that the use of the high risk register for infant hearing screening identifies only 50 % of infants with a significant hearing loss (Hayes & Northern,

1996). The mission has been to establish a screening method with a higher rate of identification. This is an admittedly daunting task for everyone involved in the process of screening infant hearing. The use of a high risk register as an initial screening technique, with those who fail being given an auditory brainstem response (ABR) screening test, has been used to make the task of universal screening slightly more manageable. The advent of otoacoustic emissions (OAEs) has prompted the idea of universal screenings, in which every infant born will be screened for hearing loss before leaving the hospital.

The prevalence of known etiologies for hearing loss has been shifting over the last 50 years. Technology has improved at a rapid rate in recent decades, bringing not only new methods of testing, but also new methods of treating very sick infants, and advances in identifying previously unknown causes of hearing loss, particularly in the area of genetics. Thus, there are currently causes of hearing loss which were previously common, but which now rarely occur, such as RH incompatibility and rubella. Additionally, more premature infants with younger EGAs are surviving. Associated issues such as maternal drug use, hypoxia, and intraventricular hemorrhage are factors that have not been fully examined as they relate to hearing impairment. Another issue which is in need of further examination in relation to hearing impairment is the presence of noise in the NICU and its possible synergistic affect with other risk factors.

Thus, there is an ever present need to update the effectiveness of the high risk register. Additionally, the high risk register is useful to determine follow-up plans, particularly in the case of those risk factors associated with late onset or progressive hearing loss, such as cytomegalovirus (CMV), ECMO, meningitis, and ototoxic medications. Therefore, there are several issues necessitating the review of the effectiveness of the risk factors currently used as well as those not currently included.

Previous research has examined the prevalence of individual risk factors and/or the percentage of failed ABR screens for a given sample. Very few studies have looked at the

failure rate for each of the specific risk factors. Halpern, Hosford-Dunn, and Malachowski (1987) examined the correlation between risk factors present in a sample population of neonates and the presence of hearing loss as verified at a later age. Thus, the study used follow-up data for the children who had been screened with an ABR in the intensive care nursery. They found that four risk factors were correlated with hearing loss in their population: craniofacial anomalies, TORCH complex illnesses, length of stay and EGA. Kountakis, Psifidis, Chang, and Stiernberg (1997) reviewed medical records of 50 deaf and 50 normally hearing neonates to compare the risk factors present in their population with those listed by the JCIH high risk register. They found that only two JCIH risk factors, craniofacial anomalies and hyperbilirubinemia, were highly correlated with hearing loss. Additionally, they found significant differences between their population and the JCIH high risk register suggestions. Kountakis et al. (1997) found higher correlation between hearing loss and risk factors not included by the JCIH, such as length of stay in the intensive care nursery and retrolental fibroplasia (eye disorder associated with prolonged high levels of oxygen), and little or no correlation between hearing loss and risk factors which are included by the JCIH, such as low birth weight and low Apgar scores at five minutes.

The purpose of the present study was to review medical records for a large population of neonates to examine risk factor status as it relates to the result of the ABR hearing screen. In addition to the current JCIH high risk factors, length of stay, EGA, and birthweight were examined.

Method

Subjects

The medical records of 203 neonates were examined. Having an ABR hearing screening test during their stay in the neonatal intensive care unit (NICU) at St. Louis Children's Hospital (SLCH) in 1997 was a prerequisite for this study. Babies admitted to

the NICU are usually screened with an ABR prior to discharge, regardless of risk status, as long as they are 35 weeks or older post conceptional age (PCA). Of the 203 records reviewed, three records were excluded due to incomplete medical history and eight records were excluded because ABR screen was completed when the neonate was less than 35 weeks PCA. Thus, the analyzed data consisted of information from the records of 192 babies.

Procedure

Risk factors and ABR data for each neonate tested were entered into the audiology department database. Each record was assigned a number, which was used as identification throughout this study. Medical records for each patient were reviewed to determine and verify risk factor status, length of stay, EGA, and birthweight. Variables examined in this study included the 11 risk factors used by the audiology department at SLCH (see Fig. 1), as well as length of stay, birthweight in grams, and EGA.

Qualifications for risk factor status are shown on the sample referral/ history form (Figure 1). Several of the risk factors required clarification to determine presence of the risk factor. Additional considerations were as follows:

Risk Factor #

3 Congenital or perinatal infections -- only TORCH complex illnesses were included. The TORCH complex illness was counted if it was diagnosed in the chart, listed on the discharge summary, or if it was noted that the child tested positive. If the mother had a history of one of these illnesses, but the child's status was not determined, the child was not considered to qualify for this risk factor.

4 Bacterial meningitis -- if the final diagnosis was "probable meningitis," meningitis was counted as a risk factor.

6 Hyperbilirubinemia -- only those neonates given an exchange transfusion for hyperbilirubinemia were counted.

7 Family history of hearing loss was the most difficult risk factor to verify. It is not information that is routinely requested of all parents.

9 ECMO or PPH -- this risk factor was counted if one of these was specifically noted in the chart.

11 Mechanical ventilation -- mechanical ventilation was considered to be use of an ET tube or CPAP, as well as both oscillating and conventional ventilators. Nasal cannula or blow by O_2 hoods were not included as mechanical ventilation.

5/10 Due to overlap in symptoms, the risk factors of defects of the head and neck, and stigmata or other findings associated with a syndrome known to include sensorineural hearing loss were combined. Some examples for this combined category include: diagnosis or genetic identification of Down's or other syndromes related to any type of hearing loss; hydrocephalus; low set, posteriorly rotated, deformed, or small for EGA ears; cleft lip or palate; preauricular pits or dimples; and small chin.

Two additional risk factor categories were included for the purposes of this study. Risk factor number12 was the diagnosis of otitis media in one or both ears. It should be noted that all babies were not checked for this condition. Risk factor number 13 was possible associated anomalies. This primarily involved some ambiguity relating to the possible presence of a genetic illness which is associated with hearing loss. The two main reasons for inclusion in this category were the presence of one parent as carrier for the sickle cell anemia trait, or another syndrome related to hearing loss was suspected but not officially diagnosed or ruled out at the time of discharge.

For all categories in which number of days were counted (i.e., length of stay, number of days on mechanical ventilation, etc.) the days were counted as days of life, as opposed to chronological days. Thus, 5/2 to5/7 would be equivalent to 6 days.

Results

The data gathered from the 192 neonates was analyzed in several ways. The failure rate for the total population was calculated for both a 30dBnHL and a 40 dBnHL failure criteria. For the 30dBnHL criterion, the failure rate was 24.5%, and for the 40dBnHL criterion, the failure rate was 15 %.

The data was also analyzed in relation to the number of risk factors present, and the failure rates for these groups. This data can be seen in Figure 2. This data shows that the more risk factors a group of neonates had, the smaller the sample size of that group, and the more likely they were to fail the screening. It should also be noted that there were three neonates, out of 45 with no risk factors, who failed the screening.

The frequency of occurrence of risk factors, and their failure rates were examined in two ways: 1.) the total number of neonates having the risk factor; 2.) the total number of neonates having only the risk factor specified. These results can be seen in Figure 3. The most common risk factor is ototoxic medication, followed by mechanical ventilation, low birthweight, and high Apgar scores. The most frequently failed overall is family history of hearing loss (100%), though the sample size is small (n=4). Presence of otitis media is also common, with 94% (n=18). The four most frequently occurring risk factors all have failure rates less than 50%, though this may also be due to variations in sample sizes.

Examination of individual risk factors in isolation was problematic due to small sample sizes. The most frequent risk factor occurring in isolation was ototoxic medication, which showed a 5% failure rate (N=22). The other risk factors occurred in five or fewer neonates when examined in isolation.

Along with individual risk factors, the most common combinations of risk factors were also examined. Again, with an increase in the number of risk factors, there is a decrease in the sample size, and an increase in the failure rate. For those neonates with two risk factors, they were most commonly #2 and #11, with 12% failure. With three risk factors, they were most commonly #2, #11, and #8, with 29% failure. With four risk factors, they were most commonly #2, #11, #8, and #1, with 54% failure. It may be noted that these four risk factors are the four most commonly occurring overall, as well. It is also interesting to note that risk factor #4, meningitis, was only present when there were four or more risk factors.

In addition to looking at the failures in relations to risk factors, the length of stay was also examined in relation to failure of the ABR screening. This data can be seen in Figure 4. As one might expect, as the length of stay increases, the number of neonates decreases, and the failure rate increases.

Lastly, the error rate in recording the risk factors of each child was examined. Throughout each child's stay in the NICU, prior to testing, the nurses take primary responsibility for noting which risk factors the child has. During review of the records for this study, it was determined that 84 of the 192 records reviewed did not require correction of the risk factors. Thus, 44% of the risk factor sheets were accurately filled out.

Discussion

The results for failure rate (24.5% at 30dBnHL and 15% at 40dBnHL) are in the same range as those in other studies. In 1988, Hall, Kripal, and Hepp compared failure rates of other studies and showed a range of 11.5 % to 41% at various intensity levels (30-60dBnHL). Galambos, Wilson, and Silva did a similar comparison of data from more recent studies, showing a range of 14.5% to 24.5% at 30dB. Alberti, Hyde, Riko, Corbin, and Abramovich (1983) found an 8% failure rate for a 40dB fail criterion. Differences in

failure rates may be attributed to a number of factors, including equipment and tester variability, and sample size and other population differences. In the 1994 JCIH recommendations, it was suggested that a 30dB criterion be used to identify hearing loss that will interfere with normal speech and language development. According to the results of this study, the 30dB criterion will result in a 10% increase in the number of children who fail, as compared to a 40dB criterion. This has implications for follow-up which must be taken into consideration. With the lower intensity level, one quarter (24.5%) of the children screened will fail and be sent into the system for follow-up to monitor possible hearing loss.

Very few studies have mentioned frequency of occurrence of individual risk factors. Swigonski, Shallop, Bull, and Lemons (1987) found low birth weight to occur most often. Shimizu, et al. (1990) also found low birth weight to be most common among their sample. The present study, however, found ototoxic medication to be most common. This may be attributable to differences in administration of ototoxic medication in different hospitals. The NICUs examined in the previous studies may use alternative, non-ototoxic medications to treat infections, or their administration practices may be less rigorous than those at SLCH. It is also interesting to note that TORCH complex illnesses did not constitute a common risk factor in this study, while they did in the study done by Halpern, Hosford-Dunn, and Malachowski (1987). This difference may be due to testing habits of the hospitals. It may also be due to other factors of the population, such as socioeconomic status, education, or prenatal care of the mothers involved. Regarding which risk factors were most likely to be failed, it is difficult to make comparisons or assumptions due to small sample sizes

The results of the length of stay data show that the longer a neonate is in the NICU, the more likely that neonate is to fail the screening. This finding makes sense in light of the fact that if a baby is in the NICU for a long period of time, it is because the

child is very ill, which means that there are probably more risk factors. As the data for number of risk factors shows, the more risk factors present, the more likely a child is to fail the screening. This finding also has implications for follow-up plans. The neonates who are in the NICU longer and have more risk factors are probably at greater risk for hearing loss and the need for follow-up to be certain of their hearing status is even more important.

There are also implications from the data regarding the errors in recording risk factors of neonates in the NICU. There was a 56% failure rate when it comes to accurately recording the risk factor status of a given neonate in the current study. It is clear from this data that the current method of identifying risk factor status is not thorough enough. The optimal method for identifying risk factors would include careful medical record review, physical examination of the infant, and interview with at least one parent (Hayes & Northern, 1996). One of the chief reasons for maintaining use of the high risk register is to aid in decisions about follow-up plans. If, for example, a child is listed as only having asphyxia as a risk factor, the follow-up plan may be more lenient. However, if the child actually has had a risk factor that was not noted, such as meningitis which may cause progressive or late onset hearing loss, such a hearing loss may be missed.

As is suggested by Kountakis et al. (1997), there may be differences between the risk factors listed by the JCIH recommendations and those found to be most common in a given hospital. There may be differences between hospitals in prevalence of specific disorders owing to differences in the overall patient population seen. There may also be differences in treatment which could account for variance in data found between hospitals.

In summary, the results of this study show the need for review of the high risk register that is used along with the electrophysiological hearing screening in the NICU. It is clear that the method currently used to determine risk factor status is not as accurate as it should be. The high risk register remains helpful to determine follow-up plans so that

children who may develop late-onset or progressive hearing loss will not be missed. Being aware of which risk factors are more likely to cause children who have that risk factor to fail the hearing screening can also be helpful for the audiologists and physicians determining follow-up care for the neonates involved. This is also the case with the evidence that a child who is in the NICU longer will be more likely to fail the screening.

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Figure 1a - SLCH Risk Factor Sheet

ABR Referral/History Information

Name: _____ Name & Address Parents/Guardians

MR#: _____ _____

DOB: ____ ____ Gest. Age ____ _____

35 weeks PCA on _____ _____

Discharge Date: _____ Phone # _____

Attending Physician: _____

Primary Care Physician after discharge _____ Check thos
risk factors which apply:

____ 1. Severe Asphyxia - Apgar of 0-4 at 1 minutes or 0-6 at 5 minutes.

____ 2. Ototoxic medication including but not limited to aminoglycosides, used > 5 day or in multiple courses or in combination with loop diuretics.

____ 3. Congenital or perinatal infections - TORCH complex: toxoplasmosis, rubella, cytomegalovirus, herpes simplex virus, and syphilis.

____ 4. Bacterial meningitis.

____ 5. Defects of the Head and Neck - cleft lip, cleft palate, malformed ears, low set ears, skin tags or pits.

____ 6. Hyperbilirubinemia - if levels exceed those indicating a need for an exchange transfusion.

____ 7. Family History of childhood hearing impairment.

____ 8. Low birthwight < 1.5 kg.

____ 9. ECMO and/or Primary Pulmonary Hypertension - if hyper-ventilation for: >5 days with pCO_2<30 mmHg and pH> 7.5 mmHg.

____ 10. Stigmata or other findings associated with a syndrome known to include sensorineural hearing loss (pigmentary abnormalities).

____ 11. Mechanical ventilation ≥ 5 days.

Other Pertinent Medical Information: _____

_____

Figure 1b- Risk Factors Used for This Study

	Risk Factors for Hearing Loss
Risk Factor #1	*Apgar Scores: 0-4 at 1 min., 1-6 at 5 min.*
Risk Factor #2	*Ototoxic Medications*
Risk Factor #3	*TORCH Infections*
Risk Factor #4	*Bacterial Meningitis*
Risk Factor #5/10	*Defects of Head & Neck; Stigmata of Syndrome*
Risk Factor #6	*Hyperbilirubinemia*
Risk Factor #7	*Family History*
Risk Factor #8	*Low Birthweight*
Risk Factor #9	*Mechanical Ventilation*
Risk Factor #11	*ECMO (Extra Corporeal Membranous Oxygenation)*
Risk Factor #12	*Otitis Media*
Risk Factor #13	*Possible Associated Anomalies*

Figure 2- Number of Risk Factors

	Total Number	NumberFailed
0 Risk Factors	*45*	*3*
1 Risk Factor	*36*	*6*
2 Risk Factors	*42*	*5*
3 Risk Factors	*31*	*9*
4 Risk Factors	*24*	*13*
5 Risk Factors	*10*	*7*
6 Risk Factors	*4*	*4*

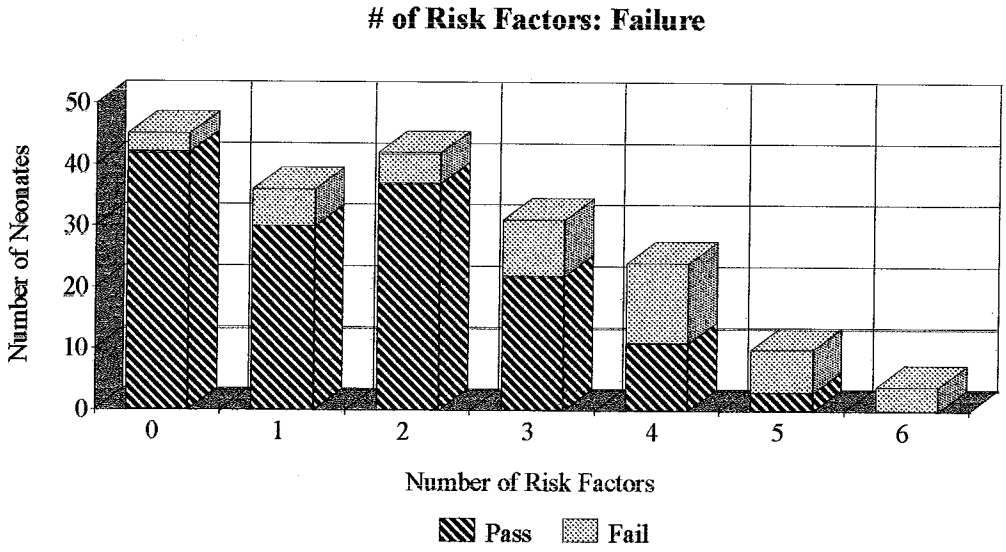
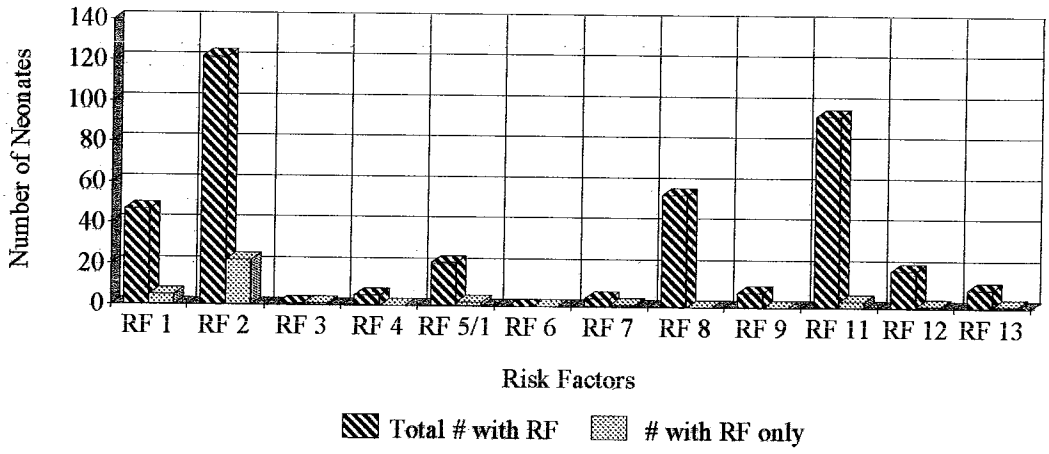


Figure 3- Risk Factors & Failure

	Total # with RF	% of sample	% fail	# with RF only	% of sample	% fail
RF 1	*47*	*25%*	*36%*	*5*	*14%*	*0%*
RF 2	*122*	*64%*	*29%*	*22*	*61%*	*5%*
RF 3	*1*	*.5%*	*0%*	*1*	*3%*	*0%*
RF 4	*5*	*3%*	*60%*	*0*	*0%*	*0%*
RF 5/10	*21*	*11%*	*52%*	*2*	*6%*	*50%*
RF 6	*0*	*0%*	*0%*	*0*	*0%*	*0%*
RF 7	*4*	*2%*	*100%*	*1*	*3%*	*100%*
RF 8	*55*	*29%*	*42%*	*0*	*0%*	*0%*
RF 9	*7*	*4%*	*29%*	*0*	*0%*	*0%*
RF 11	*94*	*50%*	*36%*	*3*	*8%*	*33%*
RF 12	*18*	*9%*	*94%*	*1*	*3%*	*100%*
RF 13	*9*	*5%*	*56%*	*1*	*3%*	*100%*

Risk Factor: Frequency of Occurrence



Risk Factors: % failure

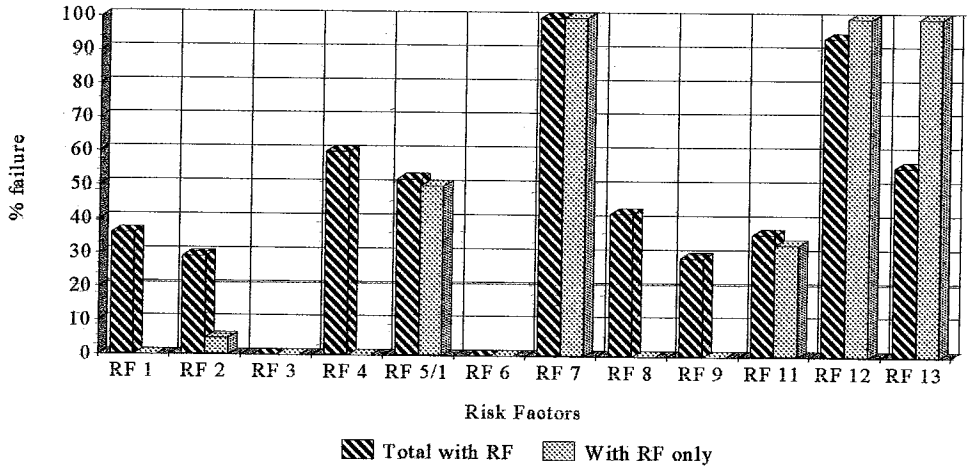


Figure 4 - Length of Stay

	<51days	51-100 days	101-150 days	151-200 days	>200 days
Passed	113	25	6	1	0
Failed	18	15	7	6	1
Total	***131***	***40***	***13***	*7*	***1***

Length of Stay: Failure